

MODALITIES OF DIAGNOSIS & MANAGEMENT OF SMALL BLUE ROUND CELL TUMOR

Dr. BRIJESH KUMAR

Senior Resident, Department of ENT, Netaji Subhash Chandra Bose Medical College, Jabalpur Madhya Pradesh, India

ABSTRACT

The small blue round cell tumour is a group of a malignant neoplasm. Sino-nasal area is a very rare site for this tumour. This case of Sino-nasal small blue round cell tumour which is reported for the first time from our hospital, which is a large tertiary care teaching institute. This tumour was found to be involving the Sino-nasal, aural & ophthalmic region. Usually, it is misdiagnosed till histopathological confirmation. Therefore, here we present a case of small blue round cell tumour of Sino-nasal region in correlation with clinical, pathological, radiological & endoscopic findings. A brief discussion on its diagnosis & modalities of management follows.

KEYWORDS: *Head Neck, Immunohistochemistry, Small Round Cell & Undifferentiated Tumours*

Received: Jul 01, 2019; **Accepted:** Jul 20, 2019; **Published:** Aug 27, 2019; **Paper Id.:** IJMPSOCT20191

INTRODUCTION

Small blue round cell tumour involving the sino-nasal, aural & ophthalmic region is being reported because of the extreme rarity of its occurrence in the ENT and head & neck region.

Usually, this is misdiagnosed till histopathological confirmation. Its correlation with clinical, pathological, radiological & endoscopic findings, diagnosis & modalities of management is being presented here.

The “small blue round cell tumours” constitute a heterogeneous group of malignant neoplasms characterized by a monotonous population of undifferentiated tumour cells with relatively small-sized nuclei and scant cytoplasm. The neoplastic elements include undifferentiated, uniform, small round to oval closely packed cells with a solitary hyperchromatic nucleus and a high nuclear-cytoplasmic ratio.

The most common neoplasms capable of presenting as a small round cell tumour are Ewing's family of tumours, rhabdomyosarcoma, neuroblastoma, lymphomas, and desmoplastic small round cell tumour.

Early and accurate diagnosis is imperative to allow a patient with a paranasal sinus or nasal cavity Small blue round cell tumours to undergo the appropriate therapy.

However, for the definitive diagnosis of a Small blue round cell tumour, light microscopic findings may be very difficult because of the frequent absence of distinguishing features. This diagnostic challenge may be further complicated if the pathologist is confronted with a biopsy of small or limited size. For proper diagnosis methods include immunohistochemical, ultrastructural, cytogenetic and molecular techniques.

Aim

- The present case report was designed to diagnose the small blue round cell tumor from sino-nasal, aural & ophthalmic area.

- Correlation with clinical, pathological, radiological & endoscopic finding.
- Management of small blue round cell tumor.

CASE REPORT

This case report is being presented from Dept. of Otorhinolaryngology & head – neck surgery, N. S. C. B. Medical College Jabalpur (M. P.).

A 16-year-old female presented with right-sided nasal obstruction & difficulty in breathing from nose for 9 months & right eye proptosis for 3 months.

On examination, patient had polypoid mass in right side nose, ear & right side eye palpebral conjunctival region. Diagnosed as right-sided nasal polyp & Nasal polypectomy with FESS was done in a private hospital but again mass developed, so ENT examination and nasal endoscopic biopsy was done in our hospital.

The standard procedure of ENT clinical examination was done -

Nose – polypoid mass right side

Eye – Right eye proptosis with chemosis & polypoid mass in palpebral conjunctival area

Face – right side generalized swelling

Ear – pinkish polypoid mass on the floor of right EAC

Posterior Rhinoscopy examination – within normal limit

Indirect Laryngoscopy examination – within normal limit

Nasal Endoscopy

Extensive necrosis & granulation in B/L nasal cavity,

Septal cartilage destroyed,

Erosion of left lacrimal bone

HPR from Nasal Endoscopic Biopsy

Undifferentiated small blue round cell tumour.

Small Blue Round Cell

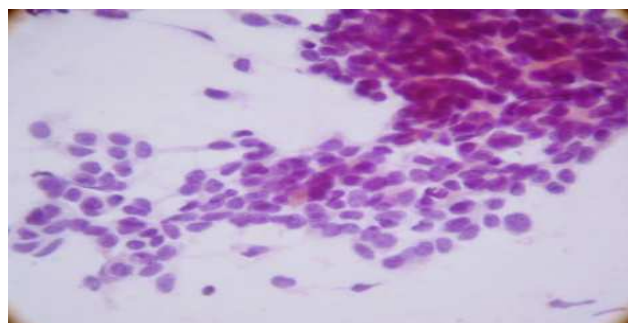


Figure 1

Immuno Histo Chemistry (IHC) Marker

- CD 99 +ve,
- Vimentin +ve,
- LCA +ve,
- Ki 67 index: 80 - 90%
- Desmin –ve,
- Synaptophysin –ve,
- Chromogranin –ve

FNAC of Upper Cervical Lymph Node

Suspicious of malignancy

Fundus

- Appears normal,
- Right-sided Retinal fold (not significant as per ophthalmologist)

Radiological Examination**USG Neck**

B/L upper cervical lymphadenopathy

CECT PNS

B/L Pansinusitis with polypoidal lesion in maxillary Ostia, nasal cavity, right exophthalmos & patchy dehiscence of medial wall of the orbit

MRI Orbit & PNS

Polypoid soft tissue in all sinuses, B/L nasal cavity, osteomeatal complex, multiple bony defects secondary to erosion on medial wall & floor of the orbit, encasing right extraocular muscle & a partially intra orbital portion of the right optic nerve, proptosis of right eyeball, intracranial extension to epidural space in right basal frontal region

X-RAY PNS and CT PNS



Figure 2

MRI ORBIT & PNS

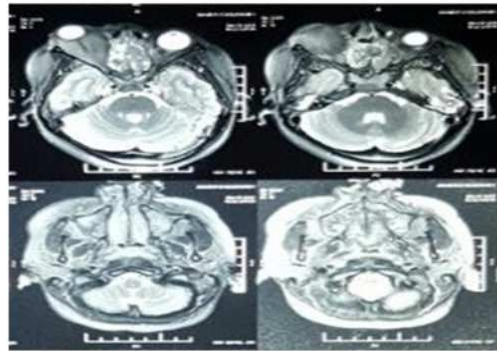


Figure 3

Pre chemo & 1st Post chemo pic

Figure 4

RESULTS

- Nasal polypectomy with FESS was done after which she got relieved from nasal obstruction but right proptosis persisted
- After getting post-op HPR of the nasal mass & FNAC report of right upper cervical lymph node, patient started on chemotherapy with (injection Ifosphamide 2gm in 200ml NS D1 to D3 & Injection cisplatin 40mg in 150ml. NS. D1 to D3.)
- The patient has clinically improved with reduced size of proptosis after receiving the first cycle of the chemotherapy and almost completely normalized after receiving the third cycle of the chemotherapy

DISCUSSIONS

Sino-nasal small blue round cell tumour is a rare malignant tumour. These neoplasms usually occurred in the sixth to seventh decades of life and rarely found in the young age group. In my case report, this tumour was found to be involving the sino-nasal, aural & ophthalmic region. Usually, it is misdiagnosed till a histopathological confirmation. The combined light microscopy, immune phenotyping and molecular findings led to the confirmation of the diagnosis of the tumors. Therefore, here I report a case of small blue round cell tumour of Sino-nasal region in correlation with clinical, pathological, radiological & endoscopic findings and its modalities of diagnosis and management.

CONCLUSIONS

- Malignancies arising in the nasal cavity and paranasal sinuses are heterogeneous. Accurate diagnosis of Sino-nasal small blue round cell tumours may be challenging. Diagnosis may be further complicated if the biopsy material is of limited quality. Advances in immune histochemistry and molecular genetics have greatly assisted in these difficult diagnoses. A precise diagnosis is important for determining aggressiveness of the tumor as well as in identifying the type of treatment needed.
- Considering all factors the treatment of small blue round cell tumour with preoperative chemotherapy followed by surgical excision of tumour & + / - radiotherapy are reliable methods of management, which gives optimal result.

REFERENCES

1. Hayes-Jordan A, Anderson PM. The diagnosis and management of desmoplastic small round cell tumor: A review. *CurrOpinOncol*. 2011; 23:385–9.
2. Abeyratna, N. I. (2013). *Current Context of Using Derivatives as Risk Management Technique of Sri Lankan Listed Companies*.
3. Barnoud R, Sabourin JC, Pasquier D, Ranchère D, Bailly C, Terrier-Lacombe MJ, et al. Immunohistochemical expression of WT1 by desmoplastic small round cell tumor: A comparative study with other small round cell tumors. *Am J SurgPathol*. 2000; 24:830–6.
4. Meis-Kindblom JM, Stenman G, Kindblom LG. Round cell tumors: differential diagnosis. *SeminDiagPathol* 1996;13(3):231–41.
5. Agarana, M. C, Bishop, S. A, & Odetunmbi, O. (2014). Optimization of banks loan portfolio management using goal programming technique. *International Journal of Research in Applied Natural and Social Sciences (IMPACT: IJRANSS)*, 2(8), 43–52.
6. Rossi S, Nascimento AG, Canal F, Dei Tos AP. Small round-cell neoplasms of soft tissues: An integrated diagnostic approach. *CurrDiagPathol* 2007; 13:150–63.
7. 44. Kim SG, Jang HS. Small cell carcinoma of the oral cavity. *J Oral MaxillofacSurg* 2001; 69:680-84.
8. Leon ME, Hou JS, Galindo LM and Garcia FU. Fine-Needle Aspiration of Adult Small-Round-Cell Tumors Studied with Flowcytometry. *DiagCytopathol* 2004; 31:147–54.
9. Ejaz A, Wenig BM. Sinonasal undifferentiated carcinoma. Clinical and pathologic features and a discussion on classification, cellular differentiation, and differential diagnosis. *AdvAnatPathol*. 2005; 12:134–143. doi: 10.1097/01.pap.0000163958.29032.56.
10. Momo Lokko, C. N, & Lokko, F. (2016). Clinical Presentation and Management of Human Ebola Virus Disease in Sub-Saharan Africa. *International Journal of Medicine and Pharmaceutical Science (IJMPS) ISSN (P)*, 2250–0049.
11. Iezzoni JC, Mills SE. “Undifferentiated” small round cell tumors of the sinonasal tract. *Am J ClinPathol*. 2005;124: S110–S121.
12. Cerilli LA, Holst VA, Brandwein MS, Stoler MH, Mills SE. Sinonasal undifferentiated carcinoma: immunohistochemical profile and lack of EBV association. *Am J SurgPathol*. 2001; 25(2): 156–63.

13. Chapman-Fredricks J, Jorda M, Gomez-Fernandez C. A limited immunohistochemical panel helps differentiate small cell epithelial malignancies of the sinonasal cavity and nasopharynx. *ApplImmunohistochemMolMorphol*. 2009; 17(3) : 207–10.
14. Yadav, S. K. S. (2016). *Customer Relationship Management Is The Need Of Today*. *BEST: International Journal of Humanities, Arts, Medicine and Science (BEST: IJHAMS)*, ISSN (P), 2348–0521.
15. Wenig BM. Undifferentiated malignant neoplasms of the sinonasal tract. *Arch Pathol Lab Med*. 2009; 133(5) : 699–712.

AUTHOR PROFILE



DR. BRIJESH KUMAR: MBBS in 2014 from Amrita School of Medicine Kochi, Kerala. MS (ENT) in 2018 from Netaji Subash Chandra Bose Medical College Jabalpur, M. P. Present time Working as Senior Resident At Netaji Subash Chandra Bose Medical College Jabalpur M. P.

Paper Publication

Trends of organ donation and awareness in Ernakulam, Kerala

Award

2nd Runner up (3rd position) in paper presentation at MPENTCON August 2018 (state conference).